Rare disease

Mediastinal germ cell tumour with massive pulmonary involvement

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Summary

Multimodality treatment, with chemotherapy and surgery, is potentially curative in case of non-seminomatous germ cell tumours. The authors present the case of a primitive mediastinal GTC with bilateral lung metastases. The patient was treated with five cycles of chemotherapy. Restaging showed reduction of the extent and of 18 FDG intake and β -HCG serum levels. The patient underwent two-step surgical excision of the tumours: mediastinal lesion and 35 lung metastases were resected by a right thoracotomy and 39 metastases were removed by a left thoracotomy. Histology showed absence of viable tumour in all the specimens. Twelve months after surgery the patient is free of disease.

BACKGROUND

Primitive mediastinal germ cell tumours (GCTs) are considered rare neoplasms, since GCTs arise prevalently from gonadal tissues. We report a case of mediastinal GCT in which bilateral pulmonary metastases were present at the time of diagnosis. A challenging treatment plan with curative intent, including two-step surgical resection, was successfully pursued.

CASE PRESENTATION

A 25-year-old male presented to emergency department with a recent history of dyspnoea after exercise and a chest x-ray showed an extensive enlargement of the mediastinal shadow with multiple bilateral coin lesions (figure 1A). CT scan confirmed a mediastinal mass with massive metastatic pulmonary involvement: 30 lesions in the right lung and 26 in the left lung (figure 1B). Positron emission tomography (PET) scan showed no extra-thoracic localisation of disease.

Percutaneous CT-guided core needle biopsy of the mediastinal lesion through a 15G Menghini needle (Hepafix, B Braun, Melsungen, Germany) resulted in the diagnosis of non-seminomatous GCT (figure 2). β -HCG serum levels was 98 mIU/ml. Clinical examination, ultrasound and MR did not demonstrate any testicular mass.

Five cycles of chemotherapy according to the scheme cisplatin, etoposide and bleomycin (PEB) were administered. CT and PET restaging showed a considerable reduction of the extent and 18FDG glucose intake of the primitive mass and lung metastases. β -HCG serum levels was 2 mIU/ml.

The patient underwent a right thoracotomy. The mediastinal mass was removed as well as 35 lung metastases. No surgical enlargement to the contiguous mediastinal structures was necessary. Histology showed no viable tumour left in the mediastinal mass and in the pulmonary nodules; only necrotic tissue was found.

One month later, the patient underwent left thoracotomy and 39 lesions were found and resected. Histology

Figure 1 Clinical presentation: chest x-ray (A) and CT scan (B).

Figure 2 Microscopic view of the histological specimen: H&E stain, original magnification 20 $\times$ (A) and 400 $\times$ (B).

Figure 3 Twelve months follow-up: CT topogram (A) and CT scan (B).

again showed the absence of viable tumour. Twelve months later β -HCG is within range (4 mIU/ml) and imaging does not show sign of relapse (figure 3).

DISCUSSION

GCTs account for 1% of all cancers and are the most common malignancy in young adult males between the ages of 15 and 35 years.¹ Even though GCTs arise prevalently (95%) from the testes and ovaries, extragonadal tissue can be primitively involved. From embryologic point of view two different origins of extragonadal GCTs can be proposed: metastases from gonadal GCTs and primary GCTs originating from migrated primordial germ cells.²

The majority occurs in the median line of the human body: sacrococcygeal region, mediastinum, pineal gland and neurohypophysis. The mediastinum is the second most common site and accounts for 15% of anterior mediastinal tumours in adults and 24% in children.²

With the advent of multimodality approach consisting of cisplatin-based chemotherapy associated with surgery, the mortality from GCT has been considerably reduced,³ whereas the incidence of GCT has been steadily increasing. This encouraging trend includes those patients with distant metastatic spread, most commonly lung.⁴ Chemotherapy can be curative in 70–80% of patients with disseminated GCTs.⁵ In this setting, surgical resection of residual masses

after chemotherapy is a well established part of treatment.⁶ The rationale lies on the inability to clinically determine whether masses contain necrosis, teratoma or viable tumour. Several attempts have been made to avoid surgical procedures: Steyerberg *et al*⁷ proposed a model to predict necrosis based on primary tumour histology (teratoma positive or negative), the prechemotherapy β -HCG serum levels, the number of residual nodules and the pathology at retroperitoneal lymph node dissection.

The presence of viable tumour after chemotherapy suggests a poor prognostic feature and indicates the need for additional anticancer treatment.⁶ In addition, elevated serum markers are considered indicative of persistent disease and are believed to be unresectable and not considered for surgical resection.⁸

Surgery of pulmonary masses after chemotherapy for non-seminomatous GCTs is currently regarded as a potentially curative intervention: patients achieving complete resection are expected to reach long-term survival, while patients with non-pulmonary metastases, persistently elevated tumours markers or viable tumour at the time of thoracotomy have a worse prognosis.⁸

In our case, the initial clinical appearance was absolutely impressive due to the massive pulmonary metastatic involvement. Nevertheless, an aggressive treatment plan with curative intent was scheduled: the patient underwent

five-cycle chemotherapy (PEB). Clinical restaging showed considerable reduction in extent of the primitive mass and lung metastases (CT scan), decreased 18FDG glucose intake (PET scan) and reduced β -HCG serum levels.

Subsequently, the patient underwent radical resection of all the pathological findings by means of two-step bilateral thoracotomy. Pathology confirmed the absence of viable neoplastic tissue in all the specimens resected and to date the patient is disease free, 12 months after surgery.

The early appearance of a massively metastatising mediastinal tumour suggests a bad prognosis end stage disease; however, the possibility of a curative treatment in such cases should always be considered.

Learning points

- ▶ GCTs primitively originating from the mediastinum are uncommon occurrences.
- ▶ Multimodality approach (including chemotherapy and surgery) is the most effective treatment in patients with non-seminomatous GCTs.
- ▶ In such patients, massive metastatising lung involvement should not contraindicate a treatment plan with curative intent.

Competing interests None.

Patient consent Obtained.

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